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of Complicated Cerebral Palsy*

Samuel Rosner, M.D.

*Mediterranean Trait
in North American Negroes*

Harry Shecter, M.D., and Abraham Frumin, M.D.

*Contributions to the Study
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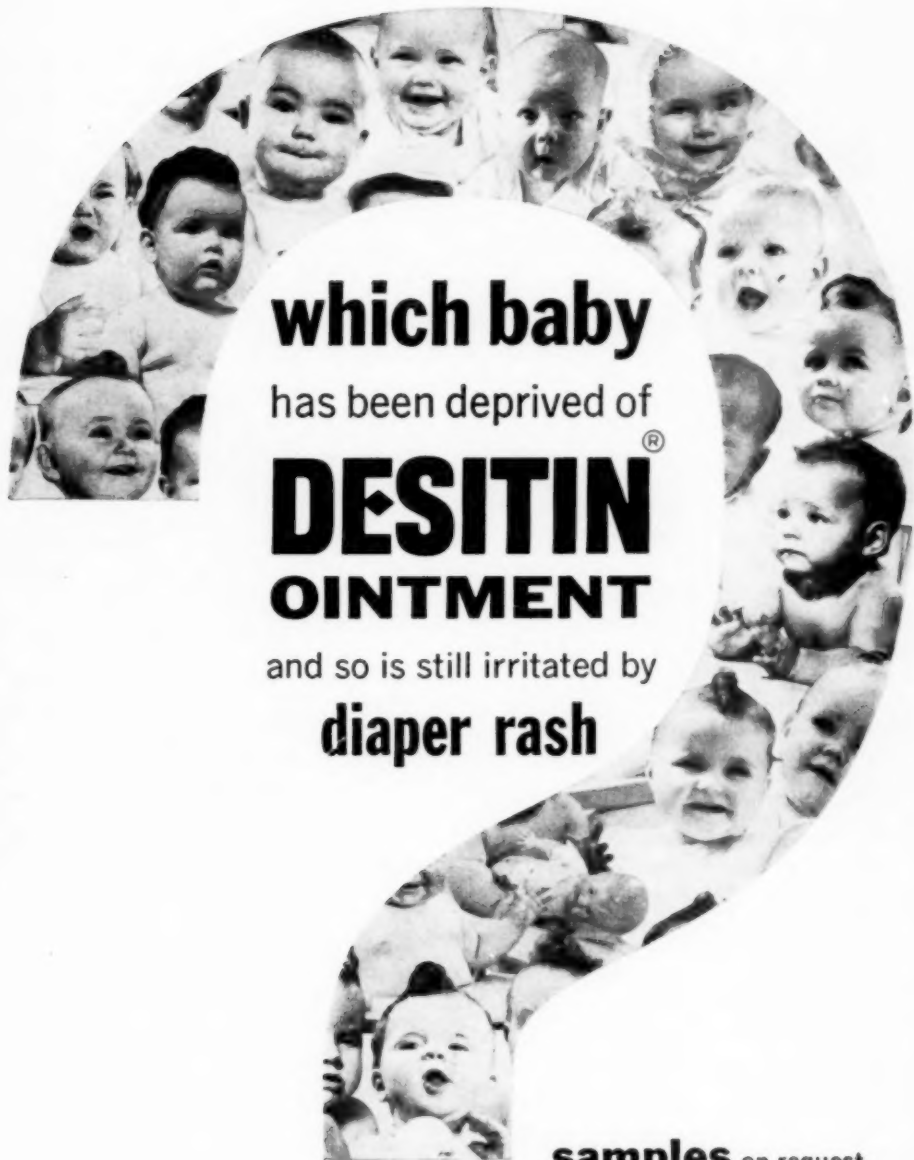
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No. 1

THE PATHOLOGY AND SURGICAL TREATMENT OF COMPLICATED CEREBRAL PALSY*

SAMUEL ROSNER, M.D., F.I.C.S.**

New York.

Cerebral birth palsy is a non-familial paralysis, paresis, or of one or more limbs which occurs before the first year of life. The palsy may be of the spastic, rigid, or choreiform athetotic type. The "complicated" patient is the one who also is suffering from mental deficiency, epilepsy, or both, in addition to the paralysis or paresis.

In fifty of fifty-two complicated cases which came to craniotomy, I found fifty patients who showed a venous or arterio-venous angioma unilaterally in the region of the lower angle of the Sylvian fissure. One child, male, aged two months showed premature closure of the fontanelles. He was suffering from grand mal seizures and spasticity in all four limbs. He has responded well to bilateral removal of bone along the coronal and sagittal sutures. There is no spasticity in his limbs and there has been no type of epileptic seizure without medication, in the fourteen months since operation. The second child who did not show a cerebral venous angioma was a complicated case of cerebral palsy. He was found to have a subarachnoid cyst of the posterior fossa with hydrocephalus. Still another child showed a unilateral cerebral venous angioma with subarachnoid cyst.

The etiology of cerebral palsy is a complex one. In this series of cases, the cause seemed to be related to birth injury by precipitate delivery, prolonged labor, instrumental delivery, haemorrhagic disease of the newborn, blood dyscrasia and possible encephalitis (one case).

Many have written on the etiology of cerebral birth palsy, among them are Little,¹ Sarah McNutt,² Beneke,³ Barnett,⁴ Capon,⁵ William Sharpe,⁶ Holland.⁷ These investigators stressed the fact that

* Under the auspices of the Department of Experimental Neurology and Neurosurgery of Stuyvesant Polyclinic, New York.

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intracranial bleeding at or before birth is a potent cause of birth paralysis. It is the author's opinion that temporary anoxia or prolonged hypoxia of the brain is the underlying cause for brain dysfunction. In previous papers,^{8,9} I had put forward the precept that unilateral venous angioma in the region of the lower angle of the Sylvian fissure is a possible cause of severe cerebral palsy with or without mental deficiency and epilepsy. The angiomata are not large and measure from one half to two inches in the greatest diameter. There are adhesions between the vessels and dura, and, the surrounding brain is cyanotic. Occasionally, one finds the middle meningeal vessels are varicose and greatly enlarged. It seems to the author that whichever of the causes for cerebral palsy prevails, that there is some bleeding which drains to the lowest point of the skull. This blood causes irritation and pressure with increase in number and size of the veins in this region so that angiomata and adhesions are formed. The drainage of blood is deficient in that oxygen cannot be adequately absorbed because of the excessive speed of transference of blood from arteries to veins, and so, hypoxia takes place.

Dr. W. B. Hamby,¹³ in his report before the First International Congress of Neurological Sciences in Brussels in July 1957, made the following pertinent observations: "The arteries supplying the lesion are inconspicuous in the gross specimen, the larger parent trunks being buried in the cerebral fissures and their branches being smaller than the more superficial dilated veins." "The complex of large veins usually lies within the subarachnoid spaces." "The identity of these vessels is impossible grossly and very difficult under the microscope."

Hamby also felt that there was a direct shunt between arterial supply and venous drainage so that there was little or no capillary bed, thereby causing cerebral anoxia. The excessive speed of passage of the blood from arteries to veins allowing insufficient time for the absorption of oxygen. I am in complete accord with the foregoing observations.

The veins involved are the middle cerebral veins, in part, and are connected by the anastomotic vein of Trolard to the superior longitudinal sinus and are connected with the inferior cerebral veins below. These veins may enter into the Basilar vein. The middle cerebral veins may open into the cavernous or sphenoparietal sinus. It is not hard to imagine that large areas of brain on

both sides of the midline may be affected by the faulty venous drainage in the Sylvian area. Of course, this is only a theory and perhaps at present, a farfetched one.

The operation was carried out on fifty-two children. Bilateral craniotomy was done in two children and four cases came to post-mortem who showed the venous angioma only on one side. One child had a posterior fossa exploration with evacuation of a subarachnoid cyst. All the others had unilateral exploration except for the child with premature closure of the fontanelles whose skull was widely opened.

The bases for operation are:

(a) The child must be a severely affected child, usually in all four limbs.

(b) He, or she, should not be responsive to any rehabilitation procedure.

(c) He, or she, should not be responsive to medication.

(d) Mental defect should not be a deterrent to surgery, provided there is no mongolism or complete amentia.

(e) Epilepsy should not be a deterrent to surgery, rather it should be an added reason for surgical intervention.

Surgical removal of the venous angioma is a fairly straight forward procedure. A three inch incision in the coronal plane centering on the pterion is brought down to bone. The skull is opened to the size of a silver dollar. The meningeal vessels are cauterized. The dura mater is incised in a stellate manner. The dura should be gently separated from the underlying structures for often adhesions to the vessels will cause profuse bleeding if lifted abruptly. At times this bleeding is unavoidable for the adhesions between the dura and the angiomata are thick and the vessel walls are exceptionally thin. The vessels very often extend to a depth of one to two inches. They should be cauterized to their depths. Sterile bismuth powder packed into the bed of the wound will help control all oozing. Then, the bismuth powder is washed out with sterile saline. The dura is left open. The temporal muscle and skin are repaired by separate silk sutures in layers.

The problem of deciding which side should be opened may be a difficult one. For, I believe that cerebral angiography^{10, 11} is dangerous in these children and so I do not use it. Pneumoencephalography is not used for it will only help us with space occupying lesions.

Clinical signs, such as ability to use the limbs on one side better than the other, differences in spasticity between one side and the other, focal epilepsy, and unilateral strabismus are aids in deciding which side to attack. Skull x-rays often will show excessive meningeal markings on the side of angioma. The electroencephalogram has been of very little help in localization of the lesion. Usually, it will show a generalized dysrhythmia with no localizing factors. One can decide on which side to operate only after weighing all the clinical and x-ray evidence. Even then one may be in a quandary. The decision, then, is to operate on the left side in a child who comes from a right-handed family.

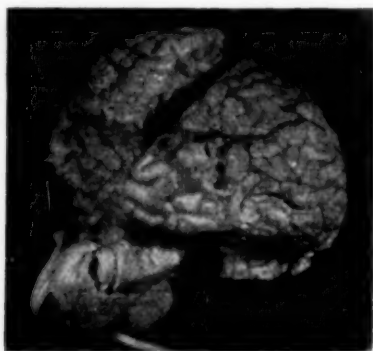


Fig. 1. Normal side.

The children showed the following clinical signs in the main:

(1) Cerebral paralysis, or paresis, of all four limbs. The limbs were spastic, rigid, or athetotic. (2) Mental deficiency in almost every case. (3) Inability to hold the head up. (4) Strabismus in one or both eyes. (5) Amblyopia. (6) Drooling of saliva. (7) Difficulty in swallowing. (8) Difficulty in respiration with stertorous breathing. (9) Epilepsy. (10) Aphasia. (11) Males showed unilateral absence of testicle in scrotum in 60 per cent of children. The sexes were almost equally divided.

Of the fifty-two patients that came to operation, nine died. One child died about a year after operation as a result of brain abscess. This child had torn open his incision, and the brain abscess resulted. Two children had died of bronchopneumonia after having returned home from hospital. Four children had died

of bronchopneumonia a few days post-operatively in hospital. One child died in a convulsive state a few months after operation. One child died at home from secondary haemorrhage.

Post-operative morbidity is usually confined to the respiratory tract with an occasional convulsion coming on in a minimal number of cases.

Two children had torn open their incisions at home. One child, mentioned above, developed brain abscess. One child healed spontaneously.

The following will show in brief outline the pertinent illustrative cases:

Case 1: L.B., male, white, age $3\frac{1}{2}$ years, delivered by low forceps, began having convulsive seizures at four months of age; well controlled by Mebaral. Electro-encephalography upon two occasions showed a petit mal type of wave. After pneumoencephalography at another hospital, the child was diagnosed as "cerebral agenesis." Left craniotomy revealed cerebral venous angioma at

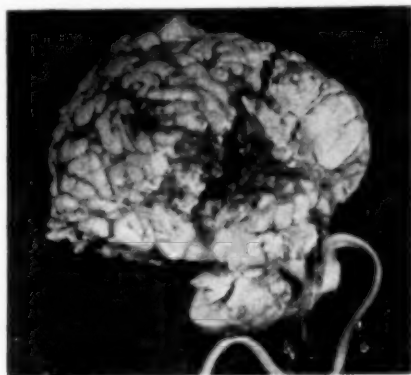


Fig. 2. Example of left sided angiona in a child with complicated cerebral palsy.

lower angle of Sylvian fissure which was cauterized to its depths, about one inch. This child could not swallow solid foods, was spastic in all four limbs with spasticity more marked on the right, he was epileptic, mentally retarded, walked with a tendency to sway to the right, could not talk and was destructive. One month after operation, the child could drink from a glass by himself,

spasticity was diminished markedly, swallowing was greatly improved. Two months after operation, the boy could walk well; was no longer destructive. Had no convulsion since operation without analeptic medication.

Case 2: P.A.A., female, white, age 2 years, 10 months. Showed strabismus at six weeks of age. She had her first convulsion at eight months. Grand mal attacks came a few times daily or once weekly. She once went five months between attacks. At ten months of age, the child was diagnosed as a cerebro-spastic. The child was mentally defective as well. She could not recognize her parents. She was aphasic, could swallow only strained foods. Rigidity was more marked on the right side.

January 7, 1954: Left craniotomy with electrocauterization of a cerebral venous angioma. Dense adhesions were separated. The cyanotic non-pulsating brain began to pulsate after cauterization of the abnormal vessels and the brain became pinkish-red in color within minutes after the destruction of the abnormal vessels.

February 20, 1954: The patient now swallows better, and holds her head better. She had one major epileptic attack. Tries to walk, strabismus is improved. Mentally the child is "brighter."

June 7, 1954: The child can crawl and stands up by herself. Plays with toys, is interested in television and is trying to talk. She has had three major epileptic attacks in five months without analeptic medication.

November 16, 1954: The child now can walk when holding her mother's hand and can say some words. Occasionally she suffers a major epileptic attack. The attacks come less frequently and are of shorter duration.

Case 3: M.A.B., male, white, age 7½ years. Normal delivery. The baby was cyanotic and was put into an oxygen tent. The doctor told the mother that the child had suffered an intracranial haemorrhage which resulted in cerebral palsy, mental defect, and major focal epileptic attacks. These convulsions began at five years of age and came on every two or three months. The child is destructive.

Left craniotomy on June 2, 1954 with cauterization of a small venous angioma at the lower angle of the Sylvian fissure.

February 24, 1954: Left facial weakness improved. Walking improved. Motor power improved. Attention and alertness improved.

October 14, 1954: The child has had two convulsions since June 1954.

August 2, 1955: The child has had no convulsions since the last visit. Has been on Mysoline.

Case 4: L.B., female, age 27 months. No pertinent obstetric history. The patient is mentally retarded, aphasic and cerebrospastic. She cannot sit up, has strabismus and showed evidence of pyknolepsy.

February 1, 1955: Left craniotomy revealed a venous angioma at the lower angle of the Sylvian fissure which was removed by electrocautery.

February 17, 1955: The child looks brighter, is more responsive to parents. Drinks from a glass.

September 27, 1955: She has more control of movements. Strabismus is improved. She responds to other children and recognizes her family. Pyknoleptic phenomena are now rare without analeptic medication. She is trying to walk and has no spasticity, can stand, and shows some response to commands.

March 10, 1956: Telephone conversation with physiotherapist shows that progress is slow and steady both physically and mentally.

Case 5: D.B., male, white, age 28 months. Prolonged labor with low forceps delivery. Mother and baby are Rh negative. Father is Rh positive. The child was cyanotic on the second day after birth and was placed in an oxygen tent and kept there for about two weeks. A pediatrician diagnosed "brain injury." Pneumoencephalography at another institution was interpreted as showing cerebral hypoplasia. At six months of age, the baby developed epilepsy. The child cannot sit up, has stertorous breathing, shows mental retardation, has oculogyria, more marked spasticity in the left limbs and is aphasic.

Right craniotomy on September 22, 1954, revealed a subarachnoid cyst with a cerebral venous angioma in the region of the lower angle of the Sylvian fissure. The angioma was cauterized and the cyst obliterated.

October 15, 1954: The child tries to lift his head, can move across the bed and tries to follow a light with his eyes.

December 31, 1954: He holds his head up better. Spasms are diminished in frequency.

February 26, 1955: He turns over and laughs. Oculogyria persists. Eyesight is poor.

September 10, 1955: Eye fixing is better. Gets head and shoulders off floor.

February 1956: Parents feel that although the child has shown definite improvement, progress is slow and that they may decide to institutionalize the child.

Case 6: J.E.G., female, age $2\frac{1}{2}$ years. Showed severe jaundice at birth. No history of obstetrical difficulty. At first, the right hand was useless but had some power at the time of examination, January 22, 1954. The child cannot sit up. "Palsy" involves all four limbs but is more marked on the right. She is aphasic and has moderate mental defect.

January 27, 1954: Left craniotomy revealed a venous angioma feeding into a central lake at the lower angle of the Sylvian fissure. These abnormal vessels were removed by electrocautery.

March 6, 1954: The child is more alert and shows no spasticity. Co-ordination is improved.

May 1954: She is beginning to crawl. Can say "bye-bye" and "Mama" and shows that she wishes to go to the bathroom.

March 1956: The father in a telephone conversation states that the child can now walk up and down stairs, is learning to speak, is alert and makes all her wants known.

My work with this small series of cases has confirmed that:

(1) The best treatment of cerebral palsy is prophylaxis. That the precepts set forth by William Sharpe⁶ are sound. He advised that any child, who was stuporous, showed difficulty in nursing, refused to nurse, showed muscle twitchings, convulsive seizures, rigidity, or cyanosis, should have spinal puncture performed. If blood was demonstrated, then spinal puncture was to be repeated every twenty-four hours until the fluid was clear. If blood was not demonstrated, then dural puncture through the anterior fontanelle or craniotomy should be done if signs persisted and if the fontanelles were bulging.

(2) Complicated cerebral palsy cases, that is, children with severe paralysis or paresis, mental defect and epilepsy are not all to be designated as hopeless. It has been my experience that almost everyone of these cases show an angioma at the level of the pterion. The removal of the angioma results in definite improvement in

motor power with markedly diminished spasticity in every case. The degree of improvement in motor power in a minority of the severe cases may be truly remarkable. The majority of cases show improved motor power, but slowly and adequately. Mentality in every case has been improved if only in small things, such as being more attentive and recognizing parents. In some children the improvement in alertness, sense of humor and making their wants known is marked. Athetotic patients do not improve as rapidly as the rigid or spastic patients. Strabismus is improved in almost every case. Difficulty in swallowing and respiration are improved in every case. The children begin to eat usual foods and no longer have stertorous breathing. Amblyopia improves slowly in most cases, but in some cases not at all. Drooling of saliva in most cases persists for a long time. Epilepsy is difficult to assess according to the scientific standard that Russell Myers¹² sets up in his admirable paper.

To the unfortunate parents of these unfortunate children scientific evaluation and standards are of little significance. They are interested in the fact that they have to cope with the epileptic attack less frequently and that the terror of its prolonged continuation no longer occurs.

Following surgical removal of the angioma, in almost every case, the epileptic attacks came less frequently and were of lesser duration. In some cases, there was no return of convulsive seizures. The shortest period of follow up was six months. The longest period of follow up was two years and three months in one case. Children that are destructive and are behavior problems usually responded poorly to surgery.

Post-mortem examination of four brains showed no gross pathology except for the operative area. The cortex on section seemed to be normal. The gyri were of normal size and the sulci were not excessively marked. Microscopic sections did not show any marked deviation from the normal in the cortex or white matter.

CONCLUSIONS

It is the author's opinion that the pathology found in these complicated cases of cerebral palsy is a definite entity, namely unilateral angioma, in the region of the lower angle of the Sylvian fissure. The hypoxia caused by these abnormal vessels may affect the brain bilaterally because of the peculiar venous anastomosis of the involved area.

After surgery, these children should have adequate physiotherapy and speech therapy for they are then in a position to benefit from these therapeutic measures.

Surgical removal of the angioma by electrocautery has resulted in definite improvement in the clinical state of children with complicated cerebral palsy so that it no longer is feasible to consider these children as hopeless.

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1882 *Grand Concourse*.

TREATMENT OF ASPHYXIA OF NEWBORN INFANTS WITH SPECIAL ATTENTION TO VALUE OF OXYGEN IN STOMACH. K. Kristoffersen. (*Nord. med.*, 57:166-171, Jan. 31, 1957). Oxygen administration in the stomach immediately after birth seems to be an ideal means for treatment of asphyxia neonatorum. It is harmless, simple, and satisfactory in preventing pulmonary complications in the first days of life, if simultaneous thorough clearing of the upper bronchial tract is carried out. Of 100 infants with asphyxia treated by this method immediately after birth, 19 died but fatal pulmonary complications occurred in only 1. No postnatal deaths were due to shock or asphyxia. These causes were found in 13 postnatal deaths among 146 infants treated by other methods.—*J.A.M.A.*

MEDITERRANEAN TRAIT IN NORTH AMERICAN NEGROES

HARRY SHECTER, M.B.Ch.B., L.M.C.C.*

and

ABRAHAM M. FRUMIN, M.D.**

Philadelphia.

Mediterranean trait in the United States most commonly occurs in people of Italian, Greek, Syrian and Armenian parentage, reaching an incidence of at least 4 per cent in Americans of Italian origin.⁷ The disorder has also been reported in North America in people of non-Mediterranean stock, such as Chinese,¹¹ Sikhs¹⁰ and Negroes.^{14, 5, 1, 6} We wish to report another instance of Mediterranean trait in a North American Negro family.

We have observed a 33 year old Negro woman (E. A.), one of ten siblings born in Alabama, and, of no known Mediterranean ancestry, to have a moderate, hypochromic, microcytic anemia which proved to be refractory to iron therapy. Examination of

TABLE 1—FAMILY A, MEDITERRANEAN TRAIT

	E.A.	R.A.	P.A.	C.A.
R.B.C.	4.8	4.30	4.01	4.16
Hb	9.4	12.6	11.7	10.5
HEMATOCRIT	35	36	34	30
M.C.V.	73	84	85	72
COLOR INDEX	0.6	0.8	0.88	0.72
RETICULOCYTES	4.4	2.2	2.8	2.8
ELECTROPHORESIS	A A	A A	A A	A A
FETAL HEMOGLOBIN	0.6	0.5	0.6	—
SERUM IRON	90	—	—	—
SERUM BILIRUBIN	0.3	—	—	—
TARGET CELLS	***	**	**	**
OVALOCYTES	**	°	°	°
STIPPLING	***	0	°	0
ANISOCYTOSIS	***	**	**	**
POIKILOCYTOSIS	***	**	**	**
Cr 51 RBC	20 days	—	—	—

the peripheral smear revealed a moderate degree of anisocytosis, poikilocytosis, hypochromia, reticulocytosis, with orthochromatic target cells, basophilic stippling, and ovalocytosis. The white cell and platelet counts were within normal limits. Examination of

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her three sons, ages 1, 4 and 6 showed similar, but less marked changes in their peripheral blood picture—only the eldest having basophilic stippling of the red cells. (Table 1).

Other investigations performed on the mother's blood included serum iron and serum bilirubin estimations, both being within normal limits; the direct and indirect Coombs' tests were negative; the erythrocyte fragility in hypotonic saline showed increased resistance with hemolysis beginning at 0.46 per cent and ending at 0.28 per cent (control 0.46 per cent—0.34 per cent) and determination of the red cell survival time by the Chromium 51 technique⁶, which showed a slight reduction in the half life survival from the normal 28-35 days, to 20 days. Filter paper electrophoresis at pH 8.6² of the hemoglobin of the mother and children showed only adult type hemoglobin to be present. Alkali denaturation¹², showed fetal type hemoglobin to be present within normal limits, being no more than 0.6 per cent (Table 1). Physical examination of the mother and children revealed no splenomegaly or any other

TABLE II—PREVIOUSLY REPORTED CASES OF
MEDITERRANEAN TRAIT IN NORTH AMERICAN NEGROES

AUTHOR	SPLENO- MEGALY	RBC	Hb	RETIC.	BILIRUBIN	NUCLEATED RBC	FETAL Hb	Hb E. PHORESIS	BONE CHANGES
SCHWARTZ et al. (Anemia)	PRESENT	4.51	9.0	5.0%	—	4	40%	A F	*
		to	to	to		to	to		to
		3.66	7.8	2.8%		18	80%	A F	***
(Trait)	ABSENT	7.0	14.7	3.0%	—	0	1.6%	A A	0
		to	to	to		to	to		
		4.63	10.5	1.6%		0	3.3%	A A	
FABER and ROTH	PRESENT	3.7	8.4	—	ICT. INDEX	73	—	—	****
STILES et al.				0.3% to 3.0%	8.0 INCR.	0	—	—	PRESENT
BANKS and SCOTT	PRESENT	MOD. ANEMIA	—	—	—	42	—	—	***
SHECTER and FRUMIN		4.01	12.6	2.2%	0.3 mg.	0	0.5	A A	0
	ABSENT	to	to	to		to	to		
		4.8	9.4	4.4%			0.6	A A	

significant findings; roentgenologic examination of the skull showed no abnormalities.

It is interesting to compare the findings in this family with the four previous reports. (Table 2).

DISCUSSION

Schwartz's cases can be divided into those showing markedly increased amounts of fetal hemoglobin by alkali denaturation and electrophoresis, and those who do not. In the former group there is erythroblastosis, splenomegaly and radiological evidence of bone changes (rarefaction and thickening)—these features, which are not present in the latter group nor in our family, are typically found in Mediterranean anemia, and help to distinguish it from the minor forms of the disease. In these cases of "anemia," in contradistinction to those without these features which may be termed "trait," the hemoglobin levels are slightly lower. In view of the marked difference in the amounts of fetal hemoglobin between the cases of anemia and those of trait, there does not appear to be any relationship to the red cell or reticulocyte count. The red cell count may be higher in anemia (Schwartz—4.51 M.) than in trait (authors—4.01 M.); the hemoglobin may be almost as low in trait (authors—9.4 gms.) as in anemia (Schwartz—9 gms.). The reticulocyte count appears unrelated to any of the other features, and may be higher in the trait (authors—4.4 per cent) than in anemia (Schwartz—2.8 per cent); the red cell survival time may be reduced in the trait. These findings are generally in agreement with Mediterranean disease in other racial groups.

The disorders most likely to obscure the diagnosis are the various hemoglobinopathies. These can only be separated from each other by paper electrophoresis, solubility of hemoglobin, etc. In Schwartz's original communication,⁹ he included two other families, and the brother of a case of Mediterranean anemia, who were later shown to be instances of hemoglobin C disease. Two cases of combined hemoglobin C—Mediterranean trait in Negroes have been described^{13, 16}, and hemoglobin G has been discovered in the blood of a Negro⁴. In other racial groups there have been recent reports of combined hemoglobin E—Mediterranean trait^{3, 16}. A point of diagnostic interest is the presence of basophilic stippling of red cells, which while not essential to the diagnosis of Mediterranean trait, has not been reported in the hemoglobinopathies.

Because of the importance of alkali denaturation and filter paper electrophoresis, the case of Faber and Roth, Stiles et al.,

and Banks and Scott cannot be unreservedly accepted as being due to Mediterranean disease. Furthermore, Stiles' case, a Negro soldier, is reported as having an Italian grandmother, and so could not truly represent the disease in a Negro. Basophilic stippling was reported in the latter, but not in the former cases.

More elaborate investigative procedures may further subdivide these disorders. The diagnosis of Mediterranean trait seems warranted in this family which is the second substantiated report of this disorder in North American Negroes.

SUMMARY

1. A Negro woman, born in Alabama, and one of ten siblings, was found to have Mediterranean trait. Study of her three children revealed a similar microcytic, hypochromic anemia with reticulocytosis.

2. Previously reported cases of Mediterranean trait in North American Negroes are reviewed.

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CONTRIBUTION TO THE STUDY OF MICROCEPHALICS

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Microcephaly is a large and important group of mental deficiency which had attracted attention not only of physicians, but also of anthropologists and psychologists. The literature on microcephaly is vast and the descriptions of the size, shape and structure of microcephalic brains are numerous and painstaking. The cause of this condition is a faulty development of the brain in utero, due to intrinsic hereditary factors or accidental intra-uterine pathologic changes such as inflammation, asphyxia, etc. Micromyelia or malformations of other systems may be associated with microcephaly. The ratio of the weight of the brain to the body normally is 1:33, but in microcephalics it may be 1:100 or more. The presence of two or more microcephalic siblings in one family points to hereditary factors in some cases. In true microcephaly, where no pathological, inflammatory or other processes are found, there is a retardation of cerebral development. In others, as was pointed out by Benda,¹ there are cystic degenerations. Almost 50 per cent of Benda's cases showed cystic degeneration of the brain. Microcephalics usually die young; the oldest in Benda's series died at 32. The exceptions are Merjeevsky's patient who died at almost 50,² Bournville's patient who committed suicide at 59,³ and Korsakov's famous Masha who died at 63.⁴

The mental life of microcephalics is greatly impoverished and most of them usually are diagnosed as idiots, but there is a transition from those whose mentality appears almost nil, who resemble decerebrate animals, to those who are active, whose movements are coordinated, facial expression pleasant and mobile, who can learn, even though with difficulty, to keep clean, to do some work, to sew, to read a little, and to play with other children.

The difference in the degree of maldevelopment, from mild to a striking reduction of size and weight, accounts for variations in clinical pictures, but intellectual weakness is not always directly proportionate to the size and weight of the brain. Cordona and

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Adriani described a microcephalic woman whose brain weighed 289 mgs. but who could dance, play cymbals and knew the names of persons and places.

The clinical diagnosis of microcephaly is warranted if the circumference of the head is less than 19 inches in an adult whose facial bones are strongly developed, while the forehead is narrow and receding, and whose x-ray of the head shows a microcephalic skull.

It seems of interest to present a group of patients whose physical, mental, psychological and radiological examinations prove them to be microcephalics, and to discuss what can be achieved by the training of these patients with extremely low mental endowment.

Case 1. B. J., white, female, born May 2, 1930 at full term by difficult instrumental delivery. Mother was 24 years of age and it was her first pregnancy. Prenatal condition of mother was said to have been good. Birth weight was 7 pounds, 4 ounces and microcephaly was noticed immediately after birth. The child began to walk at 15 months, to talk at 18 months. She had measles, chicken pox, whooping cough, mumps and scarlet fever. She was a nervous, active baby, and had a gift for mimicry. At the age of five, she was placed in a private school. The psychological test at that time showed an I. Q. of 55. She was subsequently placed in a private home for four months where she was given special treatment to overcome her neurotic behavior. Later, she began to attend a day school where she remained for two years. During this period she was tested several times. In addition to the occupational treatment, she had an extra hour of instruction daily. Her behavior was always good and she learned to write her name and to copy, but, she could not learn to count or tell time. She was cared for at home, did some housework and some sewing. She has always been neat and good-natured. She matured at 14. Menses are not regular. She was admitted to the Wrentham State School at the age of 19 with the diagnosis of microcephaly, imbecility.

The patient is a well nourished, well built, left-handed, microcephalic girl of pleasing appearance. Her height is 163.8 cms.; her weight 61 kilos.; head measurements: circumference 44.5 cms., length 15.5 cms., width 12.5 cms. Her forehead is low, narrow and receding; the top of the skull and occiput are flat. She has almond-shaped eyes with convergent strabismus (corrected

by glasses). *Laboratory examination:* Hinton, negative; blood count within normal limits. Complete urinalysis, including phenylpyruvic acid, porphyrin and sugar are negative. Fasting blood N. P. N. is 32 mg., serum total cholesterol is 252 mg., fasting blood sugar is 95 mg. per 100 cc. of blood.

X-ray examination. Lateral view of the skull showed evidence of microcephaly consistent with cranial synostosis. The hands and feet seem to be well formed.

The results of the psychological tests done prior to admission were: Chronological age 12 years, 5 months; mental age 5 years, 4 months, and an I. Q. of 43. Psychological test done on admission, revised Stanford-Binet, Form L, revealed mental age of 5 years, 2 months and an I. Q. of 32. Another test at the age of 26 revealed a mental age of 5 years, 1 month, and an I. Q. of 34, (1957). At the 4 year level she passed all but the discrimination of forms. At a 4-6 year level, she passed all but the pictures comparison. At 5 years she passed all but the counting objects. At 6 years she passed the vocabulary and the maze test.

Summary of the case:

1. Vocabulary, at 6 year level.
2. Attention span, at 5 year level.
3. Naming all the colors.
4. Reading at high kindergarten level.
5. Writing at the first grade level.
6. Learning capacity as represented by I. Q. 34, imbecile level.
7. Thinking capacity as represented by I. Q. of 34.
8. Memory for sentences is on a 5 year level and memory for digits is on a 4 year level.
9. Comprehension is at the 4 year level.

During the examination she was talkative to the point of being garrulous and tiresome, and in regard to the test questions she remarked, "they are hard for kids to answer."

During the test she asked several questions: how to do this and that, and after each test asked: "Is that all right?" She seemed to be rather apprehensive about doing things right but she was not bashful. She talks continuously, enunciates clearly, her conversation seems to be nothing more than imitation and repetition of

words heard at home and in the ward. On the whole she is rather pleasant, friendly and good-natured. She does not masturbate, controls her sphincters and is clean in her habits.

Family history. Parents are healthy, and of normal intelligence. Both are high school graduates. Father attended a university for two years. There is no consanguinity of parents. Paternal uncle is an epileptic. A maternal aunt (at 31 years of age) and an uncle (at 39 years of age) died of a hereditary disease (hematoporphyria). There is a history of pulmonary tuberculosis in the maternal aunt and uncle. The patient has two younger sisters. The second is physically and mentally healthy, married and has a normal baby. The third sibling is also microcephalic and at present is a patient at the Wrentham State Hospital. Mothers and siblings were tested for porphyria, but the tests were not conclusive.

At present, the patient is in good health. In the morning she works in the laundry, and in the industrial department in the afternoon. She attends all the entertainments. She can name all the employees in the laundry as well as the girls in the infirmary. Occasionally, she goes home with her sister for weekends and helps her mother at home. She developed epileptic seizures (of 5-7 minutes duration preceded by dizziness) in 1954, and was placed on dilantin.

Summary of the case. The patient is a well developed and well nourished microcephalic who enunciates clearly, has good motor coordination, fair comprehension, fairly good vocabulary, good table manners. She is in good contact with her surroundings, and is self-confident in everything she does or says. Her attention span and concentration are poor, she tires quickly of any mental effort, and her interest is not sustained. Her menses are irregular. Her sense of smell is poor. Her writing, reading and calculations are poor.

Case 2. B. E., white, female, born March 5th, 1942 at full term by caesarian section because of small pelvis of mother. Mother was 36 years old at the birth of the patient, and this was her third pregnancy. Birth weight was six pounds. Microcephaly was diagnosed at birth. There was no history of prenatal difficulty. She was examined at the Children's Medical Center at 5½ months because of premature closure of fontanelles and microcephaly. She walked at 15 months and talked at 2 years. She had mumps

and chicken pox. She began school at 5½ years, first in a private kindergarten for one year and in the first grade three weeks, and then excluded because she did not make progress at school, could not follow instructions, nor concentrate, and annoyed the other children.

She helped at home willingly. Her motor coordination was very good; she roller skated, jumped rope and used a scooter and a sled. She fought back when teased, otherwise she was obedient, amenable to suggestion, sociable, emotionally unstable, impulsive, good-natured, helpful at home, neat and clean. She ate and slept well. She was admitted to the Wrentham School at the age of 7½ years in 1949. The clinical diagnosis was microcephaly with developmental cranial anomalies, imbecility.

Physical examination at present shows an attractive, well-nourished, microcephalic girl who is 160 cms. tall and weighs 54 kilos. Head measurements are: circumference 43.2 cms., length 15 cms., and width 11.5 cms. She has a low, receding forehead, almond-shaped eyes with a slight convergent strabismus, large ears and winged scapulae. She bites her fingernails; talks fluently. Her blood pressure is 80/35, pulse, 80. She matured at 13, menses are irregular. Laboratory findings: urinalysis, including the test for phenylpyruvic acid and porphyrin, negative. Blood count is within normal limits. Platelets varied from 163,200 to 244,000 per cm. of blood. Hinton test, negative. Sedimentation test varied from 30 to 47 mm. at the end of an hour. X-ray showed a chronic infection in the left antrum. Blood type is A, Rh positive. Fasting blood, NPN, varied from 30-32 mgs. per 100 cc. Clotting time, 4 minutes. Bleeding time, 1 minute. Prothrombin time—whole plasma—16 seconds, diluted plasma, 42 seconds. Urinalysis, negative. Red blood count, 3,800,000, WBC, 3,200. Plasma total cholesterol is 156 mg. to 177 mg. Blood sugar 94 mg. per 100 cc. of blood.

X-ray examination. Skull showed appearance that would be consistent with microcephaly. The psychological examination on admission revealed a mental age of 4 years, and an I.Q. of 51. A test at 13 showed a mental age of 5 years, 9 months, and an I.Q. of 43. The last psychological examination at the age of 14 years and 11 months showed a mental age of 5 years, 7 months, and an I.Q. of 39. At 5 years she passed all tests but the memory

for sentences. At 6, she passed the vocabulary test, the number concepts and the maze. At 7 she passed the test for similarities.

Summary of the case:

1. Vocabulary, 6 years.
2. Attention span, 4-5 years.
3. Knowledge of colors, good.
4. Reading, first grade.
5. Writing, first grade.
6. Learning capacity represented by her I. Q. of 39.
7. Thinking capacity represented by her I. Q. of 39.
8. Memory for digits and sentences at 4 year level.
9. Comprehension from 7 year to 8 year level.

In the middle of the test she said: "I can sing for you." She sang three songs and then stopped. Her retention memory, once she learned by rote, is quite good. She is eager to do things, likes to cook. She asked the examiner for help in writing letters to her mother, to tell how much she improved (she used the word "improved" on her own initiative) and what a good girl she is. She is fond of movies and television.

Family history. Same as for sister Jane (Case #1). At present she attends the second grade. She is a great talker, is very sociable and friendly; obeys and carries out the commands during the test. She is pleasant in appearance, is tidy, well behaved, however, she is not alert.

She helps to make beds in the building, attends gymnasium classes, music class, story class, movie and all entertainments. In 1956 she had recurrent Schonlein purpura for three months, otherwise she enjoys good health. She speaks clearly and has a fairly good vocabulary as well as understanding of questions and has good motor control, good table manners, reacts normally to sensory impressions, is good-natured and self-confident. On the other hand, she has a very short attention span, poor concentration, wandering attention. She is poor in calculation and counting, her definitions are poor, her reading and writing are poor. She has a poor sense of smell discrimination; irregular menses.

Case 3. B.F.D., white male, born March 7th, 1927 at full term. Mother was 39 years old and this was her third pregnancy. Birth weight was 7 pounds, 8 ounces. Pregnancy was normal, but the

mother felt tired, having had three children in five years. At birth the child had a noticeably small head. He began to walk at 2 years and talk at 3 years. He started school at 6. He spent 2 years in the kindergarten and 2 years in the first grade, after which he entered the school for retarded children where he remained for two years. Following this he attended ungraded class for four years in a public school where he received manual training also. After school, he earned money cutting lawns and spent it on airplane models. He has a moderate defect of enunciation. He is likeable, helpful at home, able to do gardening, painting, general housework such as cleaning, etc. In school he showed very little evidence of initiative or constructive efforts. He was admitted to Wrentham State School in July 1943, at the age of 16 years 4 months, for further training.

Clinical diagnosis: Microcephaly, imbecility.

The physical examination showed a well nourished, well built adult microcephalic boy of pleasing appearance. His weight is 71½ kilos., and his height is 172½ cms. Head measurements: circumference 45.7 cm., width 12 cm., length 16 cm. He has a low slanting forehead, long nose, almond-shaped eyes and alternating strabismus. Ears are large, standing out and his ear lobules are adherent. Teeth show poor occlusion, but are in good condition. Palate is high and narrow. Coordination is good. Reflexes are hyperactive. Laboratory examination: Blood pressure: 106/54, Hinton test, negative. Serum total cholesterol is 235 mg., fasting blood sugar is 97 mg., serum NPN is 25 mg. per 100 cc. of blood. Blood type O. *X-ray examination:* the skull showed evidence of microcephaly, sella turcica within normal limits. The structure of hands and feet is normal.

Psychological examination on admission revealed a mental age of 7 years and an I.Q. of 44. Another test at 19 years and 11 months, showed a mental age of 6 years and 7 months, and an I.Q. of 44 (1957). At 5 years, he passed all tests. At 6 years, he passed all but the bead-chain test and the picture comparison. At 7 years, he passed the picture absurdities test, the copying of the diamond, and the comprehension. At 8 years, he passed the vocabulary, the verbal absurdities and the comprehension.

Summary of the case.

1. Vocabulary, 8 years.

2. Attention span, 5 years.
3. Knowledge of colors.
4. Reading and writing, second grade.
5. Learning capacity, represented by his I.Q. of 44, imbecile level.
6. Thinking capacity, represented by his I.Q. of 44, imbecile level.
7. Memory for digits, 3 years; for sentences, 5 years.
8. Comprehension, 8 years.

During the test he was cooperative and well-mannered. Answers were given promptly. He did not do well in verbal comprehension, or in rote memory. He cannot count money.

Parents are of high intellectual level. They and the other children are college graduates. There is no consanguinity of parents. Father's health is good, mother has chronic arthritis. At present, the patient works in the carpenter shop. His general health and habits are good. He is a very cooperative boy, gets along well with everyone, attends most of the entertainments and goes home on vacation. He had been very helpful at home during the summer vacation. His school behavior is good. He was reported to be very quiet and well behaved, anxious to learn, but in need of stimulation. He swims well, rides a bicycle, he is inclined to be overgenerous to others, is polite, but not as gay and carefree as the other microcephalics and he answers questions guardedly. He is right-handed. Does not know how to count money. He speaks well of his family and the personnel at school. He says that he never fights but gets mad once in a while. He claims that he has very few friends but they are good friends. He likes to watch television, does not like outdoor sports. He is quite proud of being a quiet boy.

Summary. He is well coordinated, his speech is only fair, his comprehension is good, he is good-natured, his reasoning is fairly good, his contact with his surroundings is good. He has good table manners. On the other hand, he showed poor attention span, poor concentration, poor definition, poor counting and calculation.

Case 4. S.H., white male, born January 16, 1926 at full term by difficult delivery. Mother was 47 years old and this was her first and only pregnancy, but her health was good during her

pregnancy. The patient began to walk at one year and talk at fifteen months. In school he advanced to the third grade and then was placed in a special class. He has been a member of the boy scouts and attended camp and meetings. His reading, spelling and arithmetic are at first grade level, with writing and language a little higher. He is very fond of girls, told of having three to four girl friends and added; "If I had a couple of girls to chum with I'd be all right". He has a pleasant manner and wants to work and likes companionship, but because of his amorous inclinations and mental retardation, his parents tried to keep him very busy on the farm under close supervision. The patient was admitted to Wrentham State School in December 1947 at the age of 21 years and 10 months. The clinical impression was: With other developmental anomalies—microcephaly.

The *physical examination* shows a quiet, well-behaved, well developed and well nourished, rather handsome, in spite of microcephalic head, left-handed boy, 114 cms. tall and weighing 74 kilos. He has a typical microcephalic head and large facial features. Head measurements are: circumference 47.5 cm., width 12 cm., length 16 cm. He has a low, receding forehead, almond-shaped hazel eyes with left internal strabismus and left ptosis; slight right facial paralysis; large ears, rather long uvula which deviates to the right; and hyperactive reflexes. Blood pressure, 110/80. *Laboratory examination*: Hinton test, negative; blood, group O, RH positive; urinalysis is negative, including phenylpyruvic acid. Blood count, normal. Serum total cholesterol is 278 mg., serum NPN is 26 mg., fasting blood sugar is 92 mg. (Folin-Micro).

X-ray examination shows microcephalic skull; hands and feet show normal bone and joint structure.

The psychological examination on admission revealed a mental age of 7 years, 10 months and an I.Q. of 49. During the test he was perturbed over his mistakes and proud of anything he could do correctly. Comprehension and performance were poor. Speech was rapid and forceful, superior to what would be expected with his mental age rating. He talked freely of his personal affairs. His calculation was poor and he could not understand reversing digits.

A third psychological examination at 31 was as follows: M.A. 6 years 8 months, I.Q. 44. Base was at 6 years; at 7, he passed

comprehension; at 8, he passed the vocabulary and comprehension, although he failed in all at 9, he passed the vocabulary at 10.

Discussion:

1. Vocabulary, 10 years.
2. Attention span, 5-6 years.
3. Knowledge of colors.
4. Reading, kindergarten level.
5. Writing, second grade level.
6. Learning capacity, represented by his I.Q. of 44.
7. Thinking capacity, represented by his I.Q. of 44.
8. Memory for digits, 4 years; for sentences, 5 years.
9. Comprehension, 8 years.

When, during the examination, a girl came into the room, he said: "Gee, she mixed me up". He likes movies, outdoor sports and fishing. He cannot make change or count money.

Family history: Parents are healthy, of superior intelligence and good education. There is no consanguinity of parents. The family history of both parents is negative.

At present, the patient is in good health, his habits and behavior are good. He seems eager to do things, is pleasant, polite, cooperative and smiling. He works on the farm as an assistant to the tractor driver, helps to wax floors in the building. Takes pride in his personal appearance.

Summary. His speech, articulation and enunciation are good. He is in good contact with his immediate surroundings. His disposition is pleasant. He has good table manners. His reading and writing are only fair. He is self-confident. He shows poor attention span, poor concentration, poor manner of definition, poor calculation and counting.

DISCUSSION

When these patients are presented as a group and compared in regard to their physical and mental features, background and their "trainability", the following data is obtained:

1. Two female patients are sisters.
2. All came from families of high intellectual and educational levels.
3. All were recognized as microcephalics at birth.

4. All had special training from earliest childhood, at home and in special schools.
5. All had manual training, in addition to the schooling.
6. All had almond-shaped eyes and strabismus.
7. All have pleasant cheerful dispositions and were neat in appearance.
8. All have good motor coordination, normal gait and good sphincter control.
9. All speak clearly except one (patient #3).
10. All show little evidence of initiative or constructive efforts.
11. All are poor at writing, counting or figuring.
12. All have poor elaboration of thought.
13. All have poor concentration and poor attention span.
14. They all tire easily of mental efforts; attention wanders after a few minutes.
15. All are in good contact with their environment.
16. All have learned some hand work.
17. All are unable to give the date or the time correctly. All know the place and its name, the doctor and her name (E).
18. Two of these patients (#1 and #2) have poor vision.
19. All have good hearing.
20. Two patients (#1 and #2) have poor sense of smell.
21. Two patients (#1 and #3) have poor sense of taste, (sugar, salt and pepper).
22. They all answer questions readily, but one patient (#2), interjects unrelated remarks.
23. Memory is good for the familiar things, such as home and family, etc., but poor otherwise.

They all carry non-complicated orders well, but their recollection is very poor, even of things just done or seen. They cannot repeat the story, although they laugh, if they find it funny. "You tell a silly story". However, if the same story is repeated daily for several weeks, they learn it by rote and then are able to repeat it. The impressions leave weak traces and repetition for days and weeks is necessary to remember them. They forget as easily as it is hard for them to learn. "She forgot to-morrow what she learned to-day", said the mother of one of these patients. Yet the accumulated memory of things and motions learned, made it

possible for them to learn table manners, hand work, reading and writing, however haltingly and painstakingly.

CONCLUSION

As low as their original mental endowment may be, it is apparent that they can be trained to the full extent of their limited capacities, by early home and school training. The sooner the training is begun, the better the results, as they need continuous repetition of words and actions to enable them to remember and reproduce them.

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THE RHEUMATOID FORM OF INFANTILE LEUKEMIA. P. Fornara. (*Minerva pediat.*, 7:1624-1629, Dec. 15, 1955).

Little is known about those forms of infantile leukemia the onsets of which are characterized by frank rheumatic symptoms, often leading to wrong diagnosis. The author cites histories of three children in whom the diagnosis of rheumatoid subacute leukemia was made on the basis of the roentgenograms, the findings in the bone marrow, and biopsy of the lymph nodes or other tissue. The duration of the condition was long, and fever had an undulant course. The joint manifestations were accompanied by a pseudoinflammatory involvement, which extended to the skin. Mikulicz's disease was often present concurrently. The blood findings were those of anemia with constant leukopenia. Despite blood transfusions and antibiotic, sulfonamide, and salicylate therapy, the three children died. Fornara believes that the rheumatoid symptoms of acute infantile leukemia can be attributed to the reticuloendothelial proliferative action from which the leukemia itself originates. Another Italian author, Belloni, states that the skeletal alterations that occur in the course of leukemia are the expression of stimulation of the mesenchyma by the proliferated leukemic cells.—*J. A. M. A.*

PEDIATRICS AT THE TURN OF THE CENTURY

From time to time the Archives, which was the first Children's Journal in the English language, will reprint contributions by the pioneers of the specialty over fifty years ago. It is believed that our readers will be interested in reviewing such early pediatric thought.

ANEMIA INFANTUM PSEUDOLEUKEMICA (VON JAKSCH)*

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Anemia infantum pseudoleukemica is a symptom complex, into which a certain amount of confusion has been introduced by the variety of descriptions of the condition which have crept into the literature. When Von Jaksch first described this condition in children, he thought it was a primary blood disease peculiar to these young individuals. At the time Von Jaksch published his first case, the blood in infancy and childhood had not as yet been thoroughly studied or analyzed. Since then the blood in these young individuals has been so thoroughly studied, in various conditions, that we can now see how some of the assumptions of the earlier authors were incorrect, and how confusion was thus introduced into the literature in connection with this disease.

Von Jaksch described his first case in 1889, and thought the condition which he named "anemia pseudo-leukemica infantum," was a specific primary anemia, in which there was oligocythemia, oligochromemia, a high degree of leukocytosis, a tumor of the spleen, swelling of the lymph nodes and moderate swelling of the liver. Following the description of this case, there have been published, at various times in the literature, cases either similar or corresponding in every detail to the case described by Von Jaksch. Monti, among others, undertook to specify conditions of the blood in these cases, and he characterizes the blood as being reduced in specific gravity with a diminution of the hemoglobin, a diminution of the red blood cells, a high degree of leukocytosis, and an inequality in the size of the red blood cells,

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poikilocytosis, microcythemia, and the presence of a variable number of nucleated red blood cells. These, combined with the enlargement of the spleen, moderate enlargement of the liver, Monti thought made up a specific symptom complex, primary in its nature, and easily recognizable at the bedside.

Following Monti there were cases published by Tessier, Adenoud, Glöckner, Vickery and others. These authors adhered to the original idea of Von Jaksch, that they had to deal with a primary specific condition or disease of the blood.

Fowler, in 1892, in a paper on "Splenic Anemia in Infancy," in the *British Medical Journal*, gives an analysis of a number of his cases in which the spleen was enlarged, and which in every respect, including the blood picture, corresponded quite accurately with those described by Von Jaksch. This author gives the interesting information that Gee in 1867 referred to a condition in children similar to that described by Von Jaksch, and proposed for this condition the name of lymphatic anemia.

We may say here that Wilks is another English author who proposed a similar name for this condition.

Returning to Fowler's work, he concludes that the picture of an enlarged spleen present in infancy and childhood with a definite blood picture as described by Von Jaksch, both severe and mild in form, should be grouped as primary splenic anemia.

In order to correctly understand the subsequent trend of the literature on this subject, it is necessary to state here that the original idea of Von Jaksch, that there must be a leukocytosis in all of these cases, has now been generally abandoned; for certain undoubted cases have been published which correspond quite accurately in the clinical picture to those described by Von Jaksch, which at certain periods of the disease did not show a leukocytosis.

In 1892 Fischl ventured the opinion that "anemia pseudo-leukemica infantum" was a secondary anemia, and tried to explain the confusion in the blood conditions published in connection with these cases by the fact that in infancy the microscopic picture of the blood in various forms of anemia is changeable, and showed distinctly that it was impossible to make a diagnosis from the blood picture in infancy alone—at least, in this set of cases. He described, also, severe cases of rachitis, in which the blood pictures were similar to those described by Von Jaksch,

Luzet and others, and in which autopsy showed a similar condition of the spleen—that of hyperplasia.

Epstein, also, considers this condition a secondary anemia, and the occurrence of large numbers of normoblasts and megaloblasts as evidence of mytosis of the nuclei. The presence, also, of poikilocytosis and polychromatophilia simply indicates a more or less severe disturbance of the functions of the blood-making organs depending to a degree on the condition found in the cells.

Stengel, in his article in the *Twentieth Century*, is inclined to believe that anemia pseudoleukemica infantum is an independent primary disease. In a later paper on splenic anemia, however, he is not quite so clear as to the primary nature of these conditions, and mentions the fact that the splenic enlargement in these cases overshadows the importance of the underlying original dyscrasia.

The next work of moment in connection with this condition or symptom complex is that of Wentworth. It seemed to Wentworth that "the anemia splenica infectiva" of the Italian authors, and the "anemia infantum pseudoleukemica" of the German authors and English writers were identical; that they were forms of a secondary anemia, and that they derived their peculiar symptomatology and blood pictures from the fact that this anemia occurred at an early age of life. There is very little proof, according to Wentworth, that this condition is a primary disease of the spleen or of the blood, and he also thinks there is no connection between the character of the blood and the splenic changes in these cases. Cases with identical lesions in the spleen, namely, that of chronic hyperplasia, show varying degrees of anemia. At times, as has been mentioned in this paper, the changes are marked, and at others they are not so definite. The same may be said of the leukocytosis.

Morse, in a paper on "The Relation of Chronic Enlargement of the Spleen to Anemia in Infancy," grouped a series of cases of malnutrition, some twenty in number, in which the spleen was either palpable or extended 2 or 4 centimetres below the free borders of the ribs. In two cases the spleen extended as far as the anterior superior spine and the umbilicus. The greater number of these cases were affected with a mild anemia. In only two cases did the hemoglobin fall below 20 per cent. The majority of the cases, however, were not severe anemia. Confusion is, therefore,

introduced by the attempt of Morse to group the milder and severer cases without pointing out definitely the features, especially in the clinical picture of the cases which correspond to those described by Von Jaksch. His conclusion is that the definition of Von Jaksch's disease as a severe, progressive anemia is misleading, and states that anemia is found in connection with enlarged liver and spleen in infancy, or manifestations of disturbed nutrition.

Among others who have published cases, and who follow more or less the line of thought initiated by Von Jaksch, are Baginsky, Senator, Hock, Schlessinger, Herman, Hutchison, Cozzolina, and finally, Edmund Cautley.

A departure in the consideration of the nature of Von Jaksch's disease was that made by Lehdorf. This author, after an extensive consideration of the literature, Italian, German and French, and influenced especially by the work of French writers, such as Luzet, Weil and Clerc, concludes that in all probability Von Jaksch's anemia infantum is nothing more or less than a type of myelocytic leukemia in children, and is modified in its pathological anatomy and blood picture by the complicating conditions of this period of life, syphilis and rachitis. He agrees with Luzet. Lehdorf was led to this conclusion by a careful study of a case which came under his notice and on which he made an autopsy. The blood showed megaloblasts, megalyocytes, poikilocytosis, polychromatophilia; there were nucleated red blood cells in large numbers, and he found, also, what he thought was characteristic—the presence of myelocytes in the blood with mast cells in smaller numbers. He found at all times a polynuclear leukocytosis of the neutrophilic variety, ranging from 53 per cent to 40 per cent. An examination of the organs postmortem showed a mixed marrow rich in cells; the liver cells were not changed, and there were collections of the lymphoid cells in the liver capillaries. Otherwise the spleen showed an increase of connective tissue; the pulp was rich in blood and infiltrated with large cells. There were eosinophile cells in quite a number; otherwise the changes were not marked.

Following the work of Lehdorf, which was quite extensive and, as has been said, was influenced more or less by the French school of Luzet and Clerc, we have the work of Zelenski and Cybulski. These authors, working in the clinic Jakubowski, under-

took to show, and did show quite successfully, that the presence of myelocytes was occasional in the newborn, and that in the first few weeks of life they were rarely present in the blood or absent entirely. They showed also that myelocytes in infancy and childhood could be found in a variety of diseased conditions in which anemia was present with enlarged spleen, and that they were present in from 1.5 per cent to 17 per cent of the white blood cells. Thus there is a so-called myelemia in congenital syphilis in rachitis, with enlarged spleen where there is anemia in tuberculosis, in enterocolitis, in pneumonia. Nucleated red blood cells, it may also be mentioned, occur in different conditions, and are not at all characteristic of any one condition. Of one hundred and thirty-four cases examined with reference to the presence of nucleated red blood cells, these structures occurred in thirty-six exclusive of the cases of anemia with enlarged spleen. In these same patients myelocytes occurred in eighty of the one hundred and thirty-four, but in all the cases of severe anemia with enlarged spleen these authors found normoblasts and megaloblasts. They conclude that the presence of myelocytes in the blood of infants is caused by toxins, and must be interpreted in connection with other conditions common to infancy. They have no diagnostic or prognostic significance, and are apt to be more abundant the younger the infant. The presence in large numbers in the blood of megaloblasts or gigantoblasts is significant of the fact that, though anemia is present, the processes at play in the blood were assuming a rôle of leading importance in the case.

This seems to dispose effectually of the attempt to classify conditions described by Von Jaksch and the others we have mentioned in this paper as a primary blood condition or myelemia. It may be interesting here to show that the attempt of Von Jaksch and Monti to give leading importance to leukocytosis as characteristic of these cases and of diagnostic value cannot be borne out by subsequent investigations of the blood of infancy and childhood. Geissler and Japha, in their investigations on anemia of young children, found that the average number of leukocytes in nurslings was 13,000 to the cubic millimetre, and that this number sometimes increased in the same nursling without apparent cause to 20,000 to the cubic millimetre. The average percentage of multinuclear cells was 42 per cent and they observed wide variations from 11,200 to 17,000. In other words, we should be very

careful in nurslings how we conclude as to the presence of leukocytosis, and that a polynuclear leukocytosis is a necessary accompaniment of anemia.

This investigation was especially prompted by the attempt of Monti to classify anemia of infancy and childhood into that accompanied by leukocytosis and that in which this condition was absent.

Most difficult was the interpretation of the rôle played in the various forms of anemia in infancy and childhood by the enlargement of the spleen, especially in congenital syphilis, where there is considerable enlargement of that organ and in rachitis. According to some authors there is enlargement, where others deny such an enlargement. There are, however—a fact which has been emphasized by others—children who are severely anemic who have no enlarged spleen, and in whom a thorough examination of the blood shows very little change. The same may be said of some forms of anemia with enlarged spleen.

From a consideration of what has been said as to the literature of "anemia infantum pseudoleukemica," it will be seen that there are three distinct directions of thought in regard to this condition of infancy and childhood. The first was that of Von Jaksch, who believed he had to deal with a primary blood condition accompanied by enlargement of the spleen and liver, and a definite blood picture pathognomic in its nature, which was not leukemia, but which at any time might degenerate into leukemia similar to that found in adults. These cases he called pseudoleukemic anemia infantum, because, whereas, they presented all the characteristics of leukemia, still postmortem examination showed none of the anatomical changes usually found in this condition.

The result has been a confusion of a wide-reaching character, and an encroachment on nomenclatures known for a long time to pertain to conditions found in the adult in which the blood picture is dissimilar to that found in Von Jaksch's cases in infancy. Such is the pseudoleukemia of Birch Hirschfeld.

The next trend of thought, and that which is probably the most accepted one to-day, was to consider this condition a secondary anemia due to disturbance of nutrition, which reacted on the blood-forming organs, such as the spleen and the liver, and which in turn carried with it changes in the blood due to a dis-

turbance of the functions of the blood-forming organs, but not due to a primary disease of either the spleen or the bone marrow, but rather, as intimated by the Italian authors, a disease traceable to disturbed functions of the gut reacting on other organs, and causing severe anemia. In this rubric we would classify the work of Fischl, Epstein, Wentworth.

The third trend of thought is that of the French authors, Weil and Clerc, and the German writer, Lehndorf, who consider this condition a primary anemia allied very closely to true leukemia, which was likely at any time to degenerate into this disease, and which they called myelemia. It may be stated here that no one has yet shown that any case which at the start was an anemia pseudoleukemica of Von Jaksch, or at least which presented the symptom complex, of these cases could degenerate into a true leukemia. A careful study of the whole literature fails to demonstrate such a case. One of Von Jaksch's cases, which clinically seemed to him identical with those described by him as anemia infantum pseudoleukemica, was proven postmortem to be a true leukemia, thus disproving the possibility of these two conditions merging into one another.

An analysis of the published cases of Von Jaksch's disease shows that in the blood pictures the hemoglobin varies from 10 per cent to 80 per cent in the various cases at different periods of the disease. The red blood cells vary in number from 1,000,000 to 3,000,000, or even 5,800,000 in various cases, and in 1 case, that of Fowler, the red blood cells varied from 1,000,000 to 5,000,000 to the cm.m. The white blood cells varied in the different cases from 4,800 to 5,800 to 40,000 to the cm.m. As compared to the white blood cells at one time, in the much quoted article of Lehndorf, there was a leukocytosis of 40,000 white blood cells to 1,600,000 of the red. At another time there were 15,600 white blood cells to 1,000,000 of the red.

In Morse's cases the leukocytosis in certain cases was 47,200 white blood cells as compared to 5,800,000 red blood cells. In one case there was really a leukopenia of 4,800 white to 950,000 reds.

As to the presence or absence of nucleated red cells, Von Jaksch does not give any data, inasmuch as at this time no differential counts were made. But in the cases of Lehndorf, Berggrün, Morse, Zelenski, Cybulski, the nucleated reds were present either

in small numbers or in predominant proportion. Myelocytes were present in from 1-3/10 per cent to 17 per cent of the white blood cells. As to the predominance of the lymphocytes, large or small, in some cases we have a record of the presence of 17 per cent to 41 per cent. In other cases of 11.5 per cent to 72.6 per cent. It can thus be seen that in some of these cases there is really a lymphocytic leukocytosis; and that this, in a superficial examination, might be very well confounded with a lymphocytic leukemia.

In the blood examination of nine personal cases which I consider as presenting the clinical symptoms of Von Jaksch's disease, the hemoglobin ranged from 28 per cent to 65 per cent. The red blood cells in some cases fell as low as 1,400,000, and in others as high as 4,448,000. The leukocytes ranged from 5,200 to 7,500 in one case; to 40,000 to 80,000 in another. In all cases there were nucleated red blood cells, normoblasts, megaloblasts from a number in each field to 7 per cent or 15 per cent of the red cells counted. There were several counts made in each case. In some cases at various times, the white blood cells varied in number from 11,000 to 80,000 in the cubic millimetre, with a red blood count of 2,600,000 to 3,700,000. Unless we have repeated counts in a given case, it is impossible to form a correct idea of the absolute relationship of the white to the red blood cells. Some writers have supposed that in the condition of anemia there should be a predominance of the polynuclear leukocytes. This is not so; whereas, in some cases, the polynuclear leukocytes formed 80 per cent of the white blood cells, in other cases they fell as low as 14 per cent to 15 per cent.

It has been shown that the polynuclear leukocytes varied not only in different cases as to their predominance, but in the same case there would be 45 per cent of polynuclear leukocytes at one count, and in another, taken a few days apart, there were present in 15 per cent only. So that given a leukocytosis, the predominance of the polynuclear leukocytes, as compared to the lymphocytes, is of no diagnostic value.

The myelocytes were present in all cases, varying in frequency from 1/2 per cent to 7 per cent. In some cases at different times the myelocytes varied from 1/2 per cent to 4.5 per cent in different counts. It has been shown elsewhere that the myelocytes, also, are not of specific value as differentiating these cases from

other cases of severe anemia, and the variation in the same case, at different times of the percentage of these cells, would tend to confirm this view.

The eosinophiles were present in normal percentages in all the cases.

Mast cells were present in all cases in percentages varying from 1 per cent to 4 per cent.

A study of the blood pictures in my uncomplicated cases only tends to confirm the belief expressed by others that the blood picture in this disease is not a definite pathological picture of anything but a severe anemia in subjects in whom any disturbances of the functions of the blood-forming organs causes a retrograde to the fetal structure.

A comparison of the above blood pictures with those published by Lehdorf, Fowler, Monti and Berggrün, Zelenski and Cybulski show a remarkable correspondence, and prove a contention which the author wishes to maintain, that though the blood picture is not specific, the clinical features of these cases are characteristic, inasmuch as so many observers agree as to the physical, clinical signs. The author has attempted to classify the pathological findings of all the published cases of Von Jaksch's disease, and in looking over the postmortem changes found in the various reports made of cases of anemia infantum pseudoleukemica of Von Jaksch we find a striking uniformity in the pathological data and a complete absence of anything suggesting true leukemia. In Von Jaksch's cases the spleen was large and firm; the liver was hard, slightly large; the mesentery lymph nodes were large, pale and firm; the heart was the seat of fatty degeneration. There were no other data given.

In Luzet's case, which, in passing, may be said to have taken the leading rôle with various authors who have written upon Von Jaksch's anemia, the spleen was very large, the capsule was thickened, there were no changes in the reticulum, although the pulp seemed to be increased. The liver showed a return to the embryonal type.

In a case published by Holt, which Wentworth thinks might be interpreted as a case of Von Jaksch's anemia, but which Holt published as a case of pernicious anemia, the spleen showed chronic hyperplasia and congestion. The heart was fatty and there were patches of chronic pneumonia in the lung.

In Adenoud's case the spleen showed hyperplasia of the follicles, increase of the pulp and congestion of the vessels.

In Glöckner's case the spleen showed an increase in the thickness of its capsule and the reticular connective tissue, the epithelioid cells were increased in the pulp, as were also the eosinophiles. There was little in the spleen to account for the changes found in the blood.

In the case published by Fischl the spleen was enlarged and the seat of chronic hyperplasia, and there were changes in the intestine due to intestinal catarrh.

In Lehdorf's case, in which the postmortem was quite complete and was carried out under the supervision of Albrecht, the marrow was rich in cells; there were normoblasts, leukocytes with granules and those without granules; there were myelocytes, eosinophiles, and giant cells, also cells containing pigment. The marrow was a richly cellular mixed marrow. The liver cells were normal; there were nucleated red blood cells in the capillaries, and myelocytes. The kidney showed parenchymatous degeneration, the heart was negative, the lungs showed peribronchitic infiltration, the spleen showed increased connective tissue, pulp rich in cells, capillaries dilated, eosinophiles present in moderate numbers, nothing abnormal found. Lehdorf was inclined, from the appearances, to regard the anatomical diagnosis of myelemia, especially supported by the appearances found in the liver and kidney, although the spleen and lymph nodes were less affected, and there was no siderosis.

In the author's personal cases the postmortem findings correspond, in a most remarkable manner, to what has been described in the cases of Von Jaksch, Luzet, Baginsky, Lehdorf, Glöckner and Adenoud.

A study of the postmortem findings of all the cases of Von Jaksch anemia published thus far, including those of the author, only emphasize the views of the Italian writers, and those of Fischl and the American writers that there is nothing pathognomic in the anatomical changes in the spleen, in the bone marrow, liver and other organs. In the spleen there has been regularly found an enlarged organ with an increase of connective tissue, a diminution of the splenic pulp, with nothing even remotely suggestive of leukemia.

In the liver there has been, in the various cases, an enlargement

due to simple parenchymatous changes, or to the fatty degeneration or simple lymphoid infiltration.

A study of the bone marrow gives at most a mixed red marrow and its constituents. There is nothing suggestive of leukemia, or a very marked disturbance in the structure, which could be designated as specific. A study of the intestine also fails to show anything but lymphoid infiltration.

The clinical picture presented by cases of anemia, by Von Jaksch, and following him by writers mentioned in this paper, is certainly easy of recognition.

The anemic habitus, the tumored abdomen, the large spleen of enormous size, the increased size of the liver, the intestinal disturbances, easily enable us to recognize such cases apart from the cases of slight anemia, with moderate enlargement of the spleen. There is nothing, however, in these cases which suggests leukemia, except it be the large liver and spleen. The course of some of these cases resulting in complete and satisfactory recovery, certainly impresses me with the fact that the condition is rather one of a severe disturbance of the nutritive functions of certain organs, such as the intestine, and its large secretive glandular system, reacting upon certain organs, such as the spleen, causing changes in the same, with secondary changes in the blood, which may assume a rôle of primary importance.

Von Jaksch's anemia is, therefore, a severe secondary anemia, with or without marked leucocytosis. Those cases which have been reported as terminating in true leukemia were really cases of leukemia from the outset. Cases of true Von Jaksch disease, if they terminate fatally, do so through some intercurrent disease, such as pneumonia or tuberculosis, to which they fall easy victims.

TETRACYCLINE AS A GROWTH FACTOR IN DYSTROPHIC CHILDREN. R. Scapaticci and G. Vaccarini. (*Minerva pediat.*, 9:61-65, Jan. 27, 1957). Tetracycline had a beneficial effect on the growth of 13 dystrophic children from 1 to 12 months of age. A dose of 10 mg. per kilogram of body weight, divided into 2 equal parts, was given daily for 15 to 30 days. A marked growth occurred during the first week of treatment. The authors believe that the drug has an effect on the hydrosaline metabolism; first the drug causes retention of water, and then it manifests its true nutritive effect on the formation and completion of new tissues. No untoward side-effects were observed.—*J.A.M.A.*

DEPARTMENT OF ABSTRACTS

Conducted by

MICHAEL A. BRESCIA, M.D., NEW YORK

MORRIS, D. and McDONALD, J. C.: FAILURE OF HYPERIMMUNE GAMMA GLOBULIN TO PREVENT WHOOPING COUGH. (*Archives Disease in Childhood*, 32:236, June 1957).

In two controlled trials fifty family contacts of whooping cough aged less than five years were injected with either gamma globulin prepared from the blood of donors hyperimmunized with a pertussis vaccine or with normal gamma globulin or an inactive inoculum (autoclaved gelatin). No evidence was found that hyperimmune G.G. reduced either the attack rate or the severity of whooping cough in the contacts. AUTHORS' SUMMARY.

VIVIANI, A. R.; BOGGIANO, R. V. and VEGA, I. O.: Intestinal Hemorrhage in a Child due to Intestinal Varices. (*La Prensa Médica Argentina*, 44:799, March 15, 1957).

The authors list some twenty-two causes of intestinal bleeding in infancy and childhood, as follows: 1. Intussusception; 2. Polyps of colon; 3. Meckel's diverticulum; 4. Volvulus; 5. Jejunal polyps; 6. Gastric or duodenal ulcers; 7. Esophageal varices; 8. Regional enteritis; 9. Gastroenterocolitis; 10. Chronic ulcerative colitis; 11. Blood discrasias; 12. Mesenteric lymphadenitis; 13. Lymphosarcoma of ileum; 14. Foreign body in esophagus; 15. Meningitis; 16. Hemorrhagic disease of the newborn; 17. Proctitis, papillitis, ulcerations and anal fissure; 18. Parasites; 19. Carcinoma of the sigmoid; 20. Intestinal tuberculosis; 21. Reduplication of bowel; 22. Bleeding of undetermined origin.

To the above causes, the authors add a case of bleeding due to varices involving the terminal end of the ileum and caecum. The patient was a ten-year-old girl who had two bouts of intestinal bleeding. The first time typhoid fever was considered, but the second episode was associated with pain in the right side of the abdomen. Physical examination and complete roentgenological study of the gastro-intestinal tract was negative. An exploratory was performed with a tentative diagnosis of Meckel's diverticu-

lum. At surgery, the terminal end of the ileum and caecum were edematous and surrounded by many varices. No cause could be found to account for the venous dilatations. Cure was obtained by resection of the involved intestine and colon. M.A.B.

PEACH, A. M. and DUDGEON, J. A.: Poliomyelitis Antibody Studies in a Group of London Children. (*British Medical Journal*, 5026:1033, May 4, 1957).

Sera from ninety London children aged 12 months to 10 years were examined for the presence of antibodies to the three types of poliomyelitis virus. Tests were carried out for both complement, fixing and neutralizing antibodies, and both methods are described. As a screening test for antibody, the neutralization method in tissue culture was found to be the more reliable guide to past infection. In the small group of children studied, subclinical infection began to take place at about the fifth year of life. The incidence of past infection with each of the three types of virus was fairly common, type 3 being slightly less common amongst the younger children. AUTHOR'S SUMMARY.

WRIGHT, S. W.: Phenylketonuria. (*Journal American Medical Association*, 165:2079, Dec. 21, 1957).

Phenylketonuria is an inborn error involving protein metabolism and characterized clinically by severe degree of mental defect, a lightening in the color of the skin, hair and eyes, and a strong aromatic odor to the urine. A bizarre pattern of behavior may be observed. The disease occurs once in every 25,000 births and is transmitted as a simple Mendelian recessive. The biochemical defect is a failure to hydroxylate the amino acid phenylalanine to form tyrosine and results in the accumulation of excessive amounts of phenylalanine in the body and in its excretion, together with its derivatives, in the urine. One of these derivatives, phenylpyruvic acid, gives a characteristic color reaction when ferric chloride or 2, 4-dinitrophenylhydrazine is added to the urine. These are simple laboratory tests and, when positive, are usually diagnostic of the disease. Emphasis is placed on the early detection of the phenylketonuric individual, since treatment with a low phenylalanine diet may be most effective in young infants.

AUTHOR'S SUMMARY.

CANESTRI, G. and FRACCHIA, C.: MENINGITIS IN EPIDEMIC PAROTITIS. (*Aggiornamenti sulle Malattie da infezione*, 3:111, March-April 1957).

The authors report on fifty cases of epidemic parotitis of which fourteen were bilateral and the remainder unilateral. Of the fifty cases, thirty-six had neurological symptoms such as headache, vomiting, nuchal rigidity, positive Brudzinski and positive Kernig. Although the remaining fourteen cases did not have any neurological symptoms, they as well as the others had abnormal spinal fluid findings indicating either a viral meningitis or encephalomeningitis. They noted a pleocytosis in all cases of 25 to 140 cells with 90 per cent lymphocytes. They also found an increase in proteins, glucose and chloride. The authors and the literature which they quote find that the virus producing epidemic parotitis produces a systemic disease and has definite neurotropic properties which in many cases is evident by the neurological findings.

M.A.B.

SWARTZWELDER, J. C.; FRYE, W. W.; MUHLEISEN, J. P.; MILLER, J. H.; LAMPERT, R.; CHAVARRIA, A. P.; ABADIE, S. H.; ANTHONY, S. O. and SAPPENFIELD, R. W.: Dithiazanine, an Effective Broad-Spectrum Anthelmintic. Results of Therapy of Trichuriasis, Strongyloidiasis, Enterobiasis, Ascariasis, and Hookworm Infection. (*Journal American Medical Association*, 165: 2063, Dec. 21, 1957).

It has been demonstrated that dithiazanine, 3-ethyl-2[5-(3-ethyl-2-benzothiazolinyldene)-1, 3-pentadienyl]-benzothiazolium iodide is an effective broad-spectrum human anthelmintic. In proper dosage, this polyvermidide is therapeutic for trichuriasis, strongyloidiasis, ascariasis, and enterobiasis. The drug also has significant anthelmintic activity against human hookworm, *Necator americanus*. It fulfills a need for an effective therapeutic for trichuriasis and strongyloidiasis. Dithiazanine is useful for the treatment of patients with either single or multiple intestinal helminth infections. It is effective for mass therapy for trichuriasis. An effective dosage schedule for the therapy of trichuriasis is 20 mg./lb. of body weight, divided into three daily doses, with a maximum of 200 mg. three times a day for five or more days depending upon the intensity and clinical severity of the infection. *Ascaris* infections usually can be eliminated with the same dosage. An almost

uniformly successful therapeutic regimen for strongyloidiasis was 200 mg. three times a day for 21 days. A dosage of 100 mg. three times a day for 5 days gave a cure rate of 100 per cent for pinworm infection.

AUTHOR'S SUMMARY.

WARSHAW, T. G.: A New Approach to the Control of Acne Vulgaris. (*New York State Journal of Medicine*, 57:3999, Dec. 15, 1957).

A clinical trial of a 20 per cent aqueous solution of aluminium hydroxychloride produced evidence in thirty-two of thirty-three acne vulgaris patients of decreased sebaceous gland activity with clearing of the initial complaint and with good maintenance of the desired result. A control group of thirty-three acne patients showed less rapid improvement of the acne vulgaris condition. There were no untoward effects, but there was an occasional deposit of salt. More important, a new avenue of reasoning in the therapy of acne vulgaris seems worthy of further investigation. The inhibition of eccrine sweating seems to be a satisfactory means of decreasing sebaceous gland overactivity, a mechanism in the production of acne vulgaris.

AUTHOR'S SUMMARY.

KATZ, P.: Behaviour Problems in Juvenile Diabetics. (*Canadian Medical Association Journal*, 76:738, May 1, 1957).

The diabetic status of a patient may vary with his emotional state because of: (A) Alterations by the patient of his therapeutic regimen. (B) Metabolic reactions to situational life stresses. It, therefore, behooves a physician to be concerned with the emotional problems of a juvenile diabetic. The major etiological factors in the high incidence of behaviour problems in juvenile diabetics seem to be: (A) An unsatisfactory home situation, made worse by the presence of a diabetic child. (B) The regimentation of the patient's life necessitated by the current therapy of diabetes. Forty-two per cent of all juvenile diabetics seen by the staff at the Children's Hospital of Winnipeg over a 6-year period recently had behaviour problems. A physician who assumes the management of a juvenile diabetic must devote time and effort toward establishing a good physician-patient relationship.

AUTHOR'S SUMMARY.

BOOK REVIEWS

Conducted by

MICHAEL A. BRESCIA, M.D., NEW YORK

FOR FUTURE DOCTORS. By Alan Gregg, M.D. Cloth. Pp. 165. Price \$3.50. Chicago, Ill.: The University of Chicago Press, 1957.

This book contains eleven essays by the late Alan Gregg which were delivered as addresses to as many distinguished groups. These essays are literary gems and contain most sagacious advice and council. After reading these essays one feels the urge to speak to and question the writer. It reminds me of Osler's series of addresses which were published under the title of "Acquaintance with Other Addresses," and was given to me and my classmates by Eli Lilly and Company as a gift on graduation.—*M.A.B.*

SICK CHILDREN. DIAGNOSIS AND TREATMENT. By Donald Paterson, M.D., F.R.C.P. and Reginald Lightwood, M.D., F.R.C.P., D.P.H. 7th Ed. Cloth. Pp. 593. Illustrated. Price \$8.75. Philadelphia, Pa.: J. B. Lippincott Company, 1956.

This is a fairly comprehensive but not exhaustive text on pediatrics. The many conditions commonly seen in this specialty are briefly and succinctly presented which makes this a more valuable text for the beginner in the field of pediatrics. A chapter devoted to the diseases of children in the tropics is of interest, and is not usually included in texts by American authors. Some of the illustrations are quite familiar but others are new additions to the pediatric gallery. A chart on ossification data at the end of the book is the clearest, and best seen by this reviewer.

The only criticism in an otherwise fine text was that the pages devoted to the index were not placed in order. *M.A.B.*

ITEMS

A STUDY OF *TINEA CAPITIS*. (New Zealand M.J., 53:162-165, April, 1954). A survey of school children in the Auckland metropolitan area revealed that the incidence of tinea capitis is uncommon in the summer months, there being only one case found by Wood's filtered light in a sample of 500 children between the ages of 2 and 12 years. The virulence of ringworm appears to vary somewhat from person to person even though infected from the same animal, and such lesions under the same treatment take varying times to clear up. It appears that most infections are transmitted from animal to human beings, but only rarely and less often if precautions are taken has it been proved to be transmitted from person to person. Living conditions appear to play little part in the transmission of infection, unless animals that may become infected and transmit infection to their human guardians are kept in the family. It appears that both good and poor homes are infected. In the opinion of these authors it is not necessary to keep the child from school; it appears that little or no infection is propagated among school children. Teachers and parents should be made aware of this fact, as valuable schooling is often lost when the child can least afford it.—*J.A.M.A.*

ACTH IN SYDENHAM'S CHOREA: RESULTS. (Rev. hosp. clin. 11:289-292, July-Aug. 1954). Thirteen patients with Sydenham's chorea were given corticotropin (ACTH). The patients were between the ages of 7 and 20 years. Chorea appeared for the first time in four patients and had previously appeared in annual bouts for two or three years in the rest of the patients. In the majority of the cases the electrocardiogram and other laboratory tests did not show signs of active rheumatic fever. The drug was given intravenously in daily doses of 5 mg. for a period of time that varied between 20 and 61 days. Satisfactory results were obtained in all cases, with disappearance of the choreic movements, fever, psychomotor agitation, and dysarthria within a period of time that varied between 3 and 69 days. The results are permanent up to now, two years after discontinuation of the treatment in 12 of the patients. A recurrence of moderate degree occurred in a patient five months after discontinuation of the treatment. The recurrence was also controlled by repeating the treatment. The satisfactory results in this patient remain good up to now, one and one-half years after discontinuation of the repeated treatment.—*J.A.M.A.*

KERNICTERUS FOLLOWING EXCHANGE TRANSFUSION. (*Lancet*, 1:1323-1325, June 26, 1954). Sixteen infants with hemolytic disease of the newborn were given exchange transfusions. One died during the transfusion, and kernicterus developed in four, three of whom were premature babies. Two additional cases of kernicterus occurring after exchange transfusion with packed cells in mature infants are reported. The onset of abnormal cerebral signs was preceded in the four premature babies by steadily increasing jaundice, but only in one infant were repeated estimations made of the serum bilirubin level. It was 25 mg. per 100 cc. at the time of symptoms. The diagnosis was confirmed by necropsy in one of the four premature patients, while in the other three it rested on the clinical findings. An increased serum bilirubin level that may be toxic to nerve tissue and increased vascular permeability in hemolytic infants and in nonhemolytic premature infants are considered as possible factors bearing on the production of kernicterus. The use of repeated exchange transfusions to keep the serum bilirubin levels low is mentioned.—*J.A.M.A.*

MULTIPLE RENAL LITHIASIS IN CHILDREN: FOUR CASES. (*Sem. med.*, 104:719-722, June 3, 1954). Renal lithiasis in children is rare. The author reports four cases. A boy 10 years old had an occasional mild attack of pyuria and hematuria for four years. For the last two years he complained of occasional pain in the renal region, which radiated to the back on the same side. Three weeks prior to the observation, the patient had a severe attack of acute pain, vomiting, and intestinal upset that subsided after the attack. The patient was pale and was losing weight. A roentgen examination showed a normal gallbladder and several calculi in the pelvis of the kidney. Simple radiography and excretion urography revealed 12 calculi in the pelvis of the right kidney and 3 in the lower calyx, and a greatly dilated renal pelvis. The operation consisted of a posterior pyelotomy and removal of the calculi. Antibiotics were given to the patient after the operation. No complications followed. Chemical examination of the calculi showed phosphatic lithiasis. Roentgenography one month after the operation showed no more renal calculi. The pelvis was still greatly dilated. The author believes that the calculi were formed because of stagnation of the urine caused by the dilatation of the renal pelvis due to the adhesion of the upper part of the ureter.—*J.A.M.A.*

EVERY LISTED PEDIATRIC SPECIALIST

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